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THE CUTANEOUS TUBERCULOSES IN CHILDHOOD.[1]

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The tuberculoses form part of the great class of granulomata, and, as the latter will recur constantly, it may be well to state that I understand by the term, infectious neoplasms. Childhood, for the purposes of this paper, ends with the fifteenth year.

For convenience, the subject may be divided into two classes:

I. Tuberculoses.
II. Paratuberculoses.

In the first the tubercle bacillus occurs in the tissues, in greater or less number; in the second no such finding has so far been made except in instances so rare that, for practical purposes, they are negligible, although occurring in persons who suffer from other manifestations of the disease.

I. Tuberculoses.

Four groups are usually described in this class, but a fifth must now be added. In order of frequency and importance they are:

1. Lupus vulgaris.
2. Scrofuloderma.
3. Tuberculosis verrucosa.
4. Tuberculosis vera.
5. Miliary tuberculosis.

Their symptomatology must be separately considered,[2] but etiology, pathology, prognosis, diagnosis, and treatment may, with a little ingenuity, be so arranged as to include the four varieties to be considered.

Symptomatology. *Lupus vulgaris*, in a great majority of cases, begins in childhood. The site of predilection is the face, but the disease may be disseminated over the whole integument. The primary lesion is a soft, semi-translucent nodule, deeply situated in the derma, showing through the epidermis the characteristic tint described by Hutchinson as that of apple-jelly. They may occur singly or in groups, the latter coalescing, and projecting later above the surrounding skin level. The plaque is soft, elastic and after a time begins to show a slight scaliness. Its growth is very slow, months, years even being required to form a patch one to two inches in diameter, and, as in many other instances, it is characterized by periods of quiescence and activity. Extension takes place peripherally by growth of the plaque itself or the addition of new nodules. Ulceration is often a part of the process, taking place almost invariably when the disease attacks the various orifices, nose, and mouth. This necrosis may involve all the underlying structures except bone, which is never attacked, and great deformity, such as loss of the entire tip of the nose, may result. The disease has little tendency to spontaneous healing, but during the periods of inactivity fatty degeneration and absorption of the centre of the diseased mass may occur, leaving in time a thin, atrophic, parchment-like cicatrix surrounded by a border of disease.

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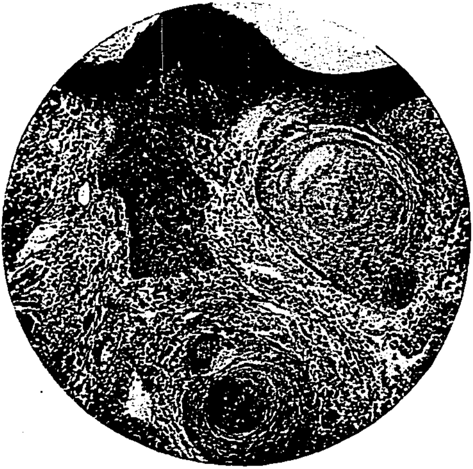
[1] Read before the Pediatric Section of the American Medical Association.

[2] The scheme of treatment of this division of the subject is a modification of that used by Dr. Bowen in Morrow's "System of Dermatology."

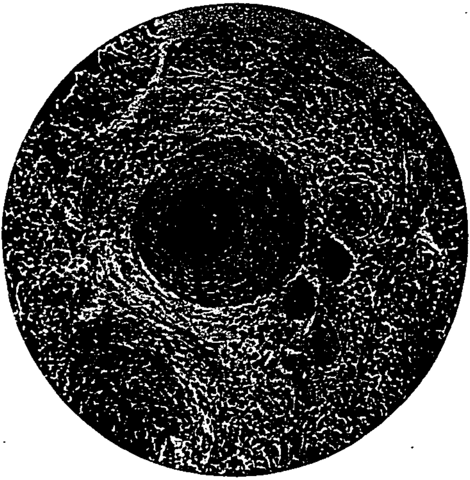
FIG. 1.



Lupus vulgaris, showing giant cells.

Accessory factors, such as position, extent, amount of infiltration, the soil and complications, give rise to variations in the type of the disease. Kaposi has applied the term *lupus disseminatus* to those cases in which the lesions are scattered, usually asymmetrically over the body. *Lupus ulcerosus* may become covered with thick, greenish crusts, and as a result of a mixed infection may fungate in the case of the nose, giving a cauliflower-like aspect. When firm œdema of the diseased tissue (lymph stasis) occurs in addition to lupus infiltration, the hypertrophic variety is seen. Its seats of predilection are the lips and buttocks. When instead or as a result of œdema the plaque becomes hard from connective-tissue increase, we have *lupus sclerosus*. *Lupus acutus* is a term applied to the disease when the process is actively extending. Papillomatous lupus has been spoken of, and verrucous disease properly belongs to another category.

FIG. 2.



Lupus vulgaris, with growth of giant cells.

On the mucous membranes, which it involves in about 30 per cent. of cases according to the figures from Doutrelepout's clinic, the lesion is papillomatous, covered with gray, exfoliating epithelium. Implication of the mucous surface is rarely primary, usually by extension from the skin. The lips and palate are favorite sites for its development. Sharplydefined ulcers, resulting from necrosis of the gray patches, are common, and cicatrization is apt to result in considerable deformity, not unlike that produced by ulcerating syphilides. It may involve the external auditory meatus, the lining of the pharynx, and the female genitalia.

Lupus may be complicated by various other tuberculous manifestations and by a systemic disturbance observed by Lespinnes (*Journ. des Mal. Cut.*, vol. iii. p. 531), very similar to that produced by injections of tuberculin. The symptoms are rise of temperature, bronchial and gastro-intestinal catarrh, and even endocarditis. It is possibly caused by toxin absorption. A grave complication of lupus in adults, epithelioma, has not been observed in children. It usually occurs in cases of long standing, and is rapidly fatal.

Scrofuloderma in modern terminology is restricted to the tuberculous lesions which develop in the subcutaneous tissue. It is usually the result of extension from neighboring foci of disease, lymphatic or osseous, and involves the skin only secondarily. It is doubtless as familiar to pediatrists as to those in dermatological practice, and I shall not stop to describe it here.

It is a long jump in point of frequency from lupus and scrofuloderma to the three remaining forms, in the adult; in children their rarity is such as to make them very nearly negligible quantities in diagnosis.

Tuberculosis vera is the name given by dermatologists to a form of tuberculous ulceration usually seen at the junction of skin and mucous membrane, as on the lips particularly, at the anus, nasal orifices, and vulva. It begins in the form of a papulo-pustule, which soon breaks down, forming a shallow ulcer with an irregular, undermined border and an uneven, grayish floor dotted with yellowish-white tubercles. It is usually covered with a thick crust. There is no tendency to healing; the process spreads peripherally until a maximum diameter of two inches is attained. The patients are usually cachectic, with evidence of tuberculosis in the internal organs. This ulceration occurs also on the mucous membranes, as of the tongue and larynx. A form of tuberculosis of the subcutaneous tissue, the scrofulous gummata (*gommes scrofuleuses* of the French), may be mentioned here, although it is more properly classed with scrofuloderma. It differs from the latter in originating in the subcutaneous tissue, not resulting from extension of neighboring foci of disease. Like scrofuloderma, it involves the skin secondarily. Tubercle formation begins in the connective tissue, the granuloma increasing in size until its diameter becomes an inch or more. The color is a livid, bluish red; the tumor is at first hard and freely movable under the skin, but later is attached to it, undergoes central necrosis, and may finally open outward, showing a deep, jagged ulceration. The gummata occur in strumous children, chiefly on the buttocks, back, thighs, and legs. Many of you have undoubtedly seen the condition as a sequel of an acute exanthem, notably measles. It occurs as a secondary development in tuberculous lymphangitis.

Tuberculosis verrucosa (McCall Anderson's scrofuloderma verrucosum) in childhood is represented in my experience by two cases, both of which were seen in the New York Hospital. In this instance the result of the activity of the tubercle bacillus is not ulceration, but the papillary outgrowth which gives it its name. The primary lesion in adults, doubtless in children also, is the post-mortem wart, *verruca necrogenica.* There may be one or several lesions, the resemblance to an ordinary wart with the addition of a dull-red areola being very striking. Spreading peripherally, the growth may attain considerable size. Three border-zones in a fully developed patch can be made out, the outer erythematous merging gradually into the healthy skin, the middle dotted with minute pustules from which pus can be expressed, and the inner consisting of the warty outgrowths. The centre behind the advancing edge is usually flattened, and finally results by absorption of the diseased mass in atrophic scarring. In the two cases referred to the three zones were not present, warty outgrowths covering the patches scattered over face, trunk, and limbs.

Miliary tuberculosis, so far as the writer's knowledge extends, has been reported in one instance only,[1] but that is of sufficient interest to warrant a short review of its points. Leichtenstein's patient, a boy, aged four years (*Münch. Med. Wochenschr.*, 1897, No. 1, p. 1) developed first a pulmonary tuberculosis which became generalized, and of which he died. In the course of the fourth week a discrete papular eruption appeared on the trunk, face, and limbs. The lesions were small, hard, red, rounded or acuminate, and clearly marked off from the surrounding skin. Successive outbreaks occurred, lasting eight to fifteen days, when the papules would disappear. The number of lesions in each relapse was never more than twelve or fourteen, but the recurrences followed each other closely. The papules occasionally became vesicular or pustular, and after resolution left a desquamating surface.

ETIOLOGY. As in other microbic infections, the seed and the soil must be taken into careful account. The tubercle bacillus is the exciting cause of these five types of disease. Their clinical features depend on the individual peculiarities of the skin and system generally of the patient, perhaps to some extent, also, on the virulence and mode of inoculation of the micro-organism.

Lupus vulgaris, as a rule, begins in childhood; but the exceptions are quite numerous, the disease appearing sometimes late in life. The reason is found probably in the greater vulnerability of the skin in early youth. Lupus progresses most rapidly then. Inoculation is usually indirect. The nodules, for example, may appear in the scars of scrofuloderma, at the borders of tuberculous fistulæ, and over diseased bones and glands. In the latter case, it sometimes happens that infection of the skin takes place through the lymphatics without involvement of the intervening tissues. Direct inoculation on tattoo marks, by ear-rings, etc., has been proved in a number of cases.

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[1] Since this was written Kaposi has reported several cases at the Twelfth International Congress.

Tuberculosis vera and verrucosa are almost invariably the result of direct inoculation. The first, with the tuberculous ulcers of the mucous membrane, is an auto-infection from secretions swarming with bacilli which pass over them from the lungs, intestines, or genital organs. Verrucous tuberculosis usually results from hetero-inoculation, though the hands may be infected, for example, from the sputa or the penis in ritual circumcision. It is seen commonly in those who handle dead, tuberculous animal matter. Its occurrence in the two children referred to is difficult to understand. I have no theory to offer.

It may be said that no conclusive proof has hitherto been adduced of an increased proportion of cutaneous tuberculosis in the offspring of diseased parents. Congenital tuberculosis is at least extremely rare. Association with pulmonary infection, whether with or without a causal relation, is an accepted fact in a proportion too large to permit its being regarded as a coincidence. Leloir found it 98 times in 312 cases; Haslund in 60 per cent.

DIAGNOSIS. The salient characteristics of lupus are its beginning in childhood, its long and tedious course, the formation near the patches and in the scars of the peculiar apple-jelly nodules. It is most frequently mistaken for syphilis, and even with the aid of the microscope diagnosis is often impossible except after long watching of the case. The history and the therapeutic test, in the writer's opinion, are practically valueless. If the tubercle bacillus is found the question is settled; but the most patient search may go unrewarded. It is often said that the bacillus does not occur once in 200 sections. Lupus erythematosus is more superficial, does not ulcerate, and does not show the nodules. Verrucous tuberculosis may be confounded with common warts, with syphilis, and eczema. The microscope will make the diagnosis with ease in the first case; its course and the clinical features enumerated, in the second; an eczema never leaves a scar, and spreads by peripheral extension, not by development of new lesions in the neighborhood. Lepra and sarcoma might be confounded with cutaneous tuberculosis. Tuberculous ulcerations are distinguished from syphilitic by their slow development, the presence of miliary tubercles and of the tubercle bacillus.

Culture experiments and the microscope furnish exact methods of diagnosis in the forms where the bacillus occurs in numbers—*i. e.*, in scrofuloderma, verrucous, ulcerative, and miliary tuberculosis, but the case is different in lupus vulgaris. Not only are the histological features very similar to those of certain superficial syphilodermata, but bacilli occur so sparsely that culture almost always fails. Reliance is best placed on the points given.

PATHOLOGICAL ANATOMY. The lesions of all the types are neoplasms of granulomatous character. Kaposi's description of the histopathology of lupus still remains the best. The nodule begins in the deeper part of the corium, and consists of cells, leucocytes, and proliferated connective-tissue corpuscles, embedded in a delicate fibrous reticulum, supplied by numerous, dilated vessels. The infiltration later becomes more dense, giant cells appear, the blood-supply is cut off by pressure, and central necrobiosis takes place. The result of this tissue death is absorption and atrophic scarring, or, in case extension has involved the epidermis, ulceration. The foci usually spread along the course of the vessels. When the epidermis is involved the prickle cells proliferate and the interpapillary projections are widened and lengthened. The boundary line disappears gradually and, by tissue loss through desquamation or suppuration, ulceration occurs. When connective-tissue proliferation takes place lupus sclerosus develops; when the papillæ enlarge a verrucose condition results; occasionally epithelial proliferation is excessive, and it is in this way that epithelioma begins. Tubercle bacilli are to be seen, but very few in number and usually within the giant cells. When found after a search through 100 sections or more, they are often single or occur in twos and threes.

In the four other varieties the pathological findings do not differ greatly from each other. The greater portion of diseased material is granulation tissue composed of leucocytes, epithelioid, and giant cells. Tubercle bacilli occur in great number in all of the lesions, from which they may easily be cultivated and inoculated. Scrofuloderma is a tuberculosis of the subcutaneous connective tissue. The tubercles coalesce, invade the corium and epidermis, ulcerating finally by cell necrosis through the latter. Tuberculosis vera is exactly the same process beginning more superficially and progressing more rapidly. The latter bears close analogy to miliary tuberculosis elsewhere.

In verrucous tuberculosis the corium is invaded by the same granulation tissue, but there is a tendency to epidermic prolifieration rather than ulceration. The papillæ are greatly elongated and broadened, with corresponding hypertrophy of the interpapillary prolongations. The surface is covered by thickened layers of horny cells which, following the dipping between the papillæ, give rise to the appearance of verrucosity. Here and there, where epidermic proliferation is greatest, may be seen minute pustules. When present they finally burst superficially, and the pits left become filled with horny masses of epidermis.

The number of tubercle bacilli is greater than that in lupus, less than in tuberculosis vera.

In the single case of miliary tuberculosis the papules were found to consist of tubercles situated in the papillary layer and containing numerous bacilli. A secondary infection by pus-organisms accounted for the pustulation.

PROGNOSIS. This is more or less unfavorable in all forms. In tuberculosis vera and miliaris the danger is to life itself, since they may justly be regarded as part of a grave systemic infection. All forms are rebellious to treatment, prone to relapse, and show no tendency to spontaneous healing. A scar is, moreover, the invariable result with the best of treatment. Left untreated, the disease may continue and progress indefinitely. Prognosis is modified by the variety: lupus is much more difficult to cure than scrofuloderma or verrucous tuberculosis. Possibility of general infection from the skin lesion should always be borne in mind.

TREATMENT. Specific treatment in the shape of tuberculin may be dismissed, it may be said, with little fear of contradiction, as a complete failure at present. I have never seen the remotest good result from its use. Prophylaxis should not be neglected. Those who have a tendency to the development of tuberculosis should be hedged about by every hygienic precaution; the skin should not be exposed to contact with the virus, and should be antiseptically cleansed when so exposed; it should be protected when possible by destruction of underlying disease. There is no need to dilate upon the necessity for general hygienic and therapeutic measures. These are the same and equally important in the skin as in other organs.

Chief reliance must, after all, be placed on local treatment. It may be summed up in one word—removal. The means to that end are limited only by the bounds of human ingenuity. The ideal method, advocated by Lang, of Vienna, is *excision* and grafting; but, unfortunately, this is limited to small areas, and the disease is apt to return even then. *Curetting* with a Volkmann spoon or curette offers certain advantages, notably the aid to the operator, in the feeling of resistance given by healthy tissue. It is most useful in large areas, and should then be done under general anæsthesia. *Linear scarification* may be done with local anæsthetics by means of a single blade or a scarificator consisting of several parallel cutting edges. While not curative in itself, this method offers advantages as an adjunct to other treatment. The scarifications are done in parallel lines and crossed at right angles. It is useful in locations, lips or ears, difficult of access to other means. A further advantage is that it produces a soft, supple cicatrix; objections to it are the long duration of treatment and possibility, in common with other surgical methods, of general infection through lymph channels. It is particularly applicable in lupus sclerosus. *Cauterization* may be done with the Paquelin or electro-cautery, or by chemicals. The first is useful in large areas; the second, in picking out single nodules deep in the cutis or recurring in a cicatrix. *Chemical cauterization* is a method not in great vogue in this country. Arsenical pastes, Vienna or Marsden's, destroy the diseased parts, but should not be used over areas greater than one and a half inches square. They are excessively painful. Silver nitrate in solution or solid may leave a permanent stain in the skin. It is impossible to limit the action of zinc chloride; salicylic acid is useful only to remove thickened epidermis; carbolic acid has no value. Pyrogallic acid (paste, 10 per cent.) acts only upon foci of disease. *Electrolysis* may be used to destroy isolated lesions.

Success in treatment is largely a question of the skill shown in selection and carrying out of the method. No single mode can be followed successfully, and often resort must be taken to all in one case before success is assured. For example, a large ulcerated plaque of lupus on the cheek or tuberculosis verrucosa on the hand may be curetted thoroughly and the actual cautery applied to the pockets of disease. The wound may be dressed antiseptically or covered with a layer of pyrogallic ointment (there is danger of intoxication from the latter if too large a surface be attacked). The acid destroys disease which has escaped the scraping and cauterization. When a clean surface is secured it is allowed to granulate, plasters (mercury) or the solid stick restraining exuberant granulation. After the scar has formed it may be scarified to make it soft, and recurrences may be destroyed by electrocautery or electrolysis. Fortunately, we have now at hand a means of reducing the hideous, puckered cicatrices of cutaneous tuberculosis in thiosinamin. A 10 per cent. solution in glycerin and water is injected hypodermatically every second or third day in 10-minim doses.

II. PARATUBERCULOSES.

In 1894 Fournier published in Paris a book which he called *Les Affections Parasyphilitiques.* By the term he means affections not necessarily syphilitic in themselves, but developing and flourishing upon a specific basis. The step is not far from syphilis to tuberculosis in pathology, and by analogy it is not strange that there should be a crowd of diseases with a thoroughly recognized relationship to the latter, in spite of the fact that Koch's bacillus is only very rarely found in their lesions, even then probably resulting from accidental contamination. Many efforts have been made of late to find and fix a place for the paratuberculoses, notably during the Congress of Dermatologists last year in London, by Hyde, Hallopeau, Crocker, and others. (The name I have used was not suggested—in fact, none was given—but it seems to fit the case.)

Very few dermatologists are ready to accept as corollaries to tuberculosis all the diseases so designated by Hyde and Hallopeau. Tommasoli appears to have the right of the question when he says that it is a mistake to consider that dermatoses, such as Hebra's pityriasis rubra, general ichthyosis, dermatitis exfoliativa, eczema even, have any causal relationship with tuberculosis other than that, in the impairment of the resisting powers following a long-standing disease of this kind, the bacillus finds its entrance easier and its soil more favorable. Upon one point, although the guess is at present a hazardous one, opinions, in a majority of observers, are apparently agreed, viz., that the origin of the diseases commonly accepted as paratubercular is toxinic. They are, in all likelihood, due to the action of poisons derived from distant foci of disease.

For convenience, those diseases concerning whose title to a place here there is little dispute may be ranged in three subdivisions: (1) the scrofulides; (2) the tuberculides; and (3) hyperchromia. The scrofulides, first described by Duhring, are pyodermic affections which, if toxinic in origin, are so probably in the sense that pus organisms are more easily inoculated upon a skin weakened in resisting power by circulating poisons. In this scheme they include:

a. The small pustular scrofulide.
b. The large pustular scrofulide.
c. Certain suppurative folliculites.

The second subdivision, called tuberculides by Darier, includes, in his classification, a number of affections which may be narrowed to

a. Lichen scrofulosorum.
b. Acne cachecticorum.
c. Erythema induratum scrofulosorum.
d. Grouped folliculitis.
e. Possibly the disease usually described as hydradenitis.
f. Lupus pernio.

The first three are typical paratuberculoses. Kaposi says one of the diagnostic points in lichen scrofulosorum is its occurrence in the tuberculous. The origin of *d* and *e* is more than doubtful. None of these diseases are commonly pustular, but they may become so by accidental contamination. In this way they are separated from the scrofulides, which are pure pyodermias.

The last class includes but one affection, a hyperpigmentation, a "pigmentary dysgenesis" common to various cachexias, of syphilis, cancer, paludism, alcoholism, diabetes, as well as of tuberculosis. In this instance it may aptly be called the pigmentary tuberculide in analogy with the pigmentary syphilide. Fournier (*Les Affections Parasyphilitiques*) describes a case[1] in a young girl, which could be distinguished from the specific dyschromia only with the greatest difficulty. The pigment is derived from the blood originally, but its chemical composition is changed. Once deposited in epithelial cells, for which it has a special predilection, it is practically ineradicable.

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[1] Some eighteen others have been reported by various observers.

None of the paratuberculoses are common in practice. I shall consequently describe briefly only the scrofulide, lichen scrofulosorum, and erythema induratum. Their importance lies in the fact that they are easily mistaken for syphilides, as pointed out by Hutchinson.

The members of the first group rarely occur singly in a pure state. The eruption is generally constituted of large and small pustules, frequently beginning as a perifollicular inflammation. Moreover, they are often seen complicating the tuberculides. The initial lesion may be a papule, but is usually a vesico-pustule. They may be large or small, follicular or not, grouped or irregularly scattered over the back, buttocks, legs, thighs, and arms. When large, the pustules may coalesce and become covered with thick crusts. Whether large or small a depressed cicatrix is apt to result. The course is slow and indolent. Stanley (*Brit. Journ. of Derm.*, Nov., 1893) published a case, a boy aged fourteen years, in whom the eruption was recurrent and displayed its greatest activity when tuberculosis of the cervical glands was most active. The patient progressed favorably under cod-liver oil internally and externally by inunction.

Lichen scrofulosorum is the type of the paratuberculoses. It occurs almost invariably in tuberculous patients, and very rarely does the bacillus appear in the lesions. The controversy over its presence, according to Pellizarri, arises from the fact that the toxins prepare the soil for its entrance. No improvement has been made on Hebra's original description. The eruption is extremely chronic, and consists of grouped, small, firm, flat, livid-red papules disseminated over the back, abdomen, and thighs. They may be capped by a fine scale or a tiny pustule. At first they are isolated, but become closely grouped later. The patches are not large, and may have a crescentic outline. In Austrian statistics 99 per cent. of cases showed other evidences of struma. The affection is encountered between infancy and puberty, never after the twentieth year. The papule is formed by an inflammatory infiltration about the pilo-sebaceous follicle. On involution it leaves pigmentation or faint scars.

Colcott Fox (*Brit. Journ. of Derm.*, Nov., 1895) has described several cases of a disease which he called "acne scrofulosorum in infants," and which is in all probability identical with Hebra's acne cachecticorum. His description is as follows: "The eruption is an indolent, small, papulo-pustular or acneiform one in infants, disseminated sparsely and without grouping and unaccompanied by subjective symptoms. It affects the lower limbs and buttocks, chiefly on their extensor surfaces.

The lesions appear successively or by a subacute outbreak. The papules are inflammatory, seated about a hair-follicle and acuminate in the early stage. They mostly develop a crown of pus, . . . and in involution become flattened and leave stains or scars." This is not very different from the lichen just described. In fact, as I have said, they and the pustular scrofuloderm may flourish on the same soil.

Erythema induratum (Bazin's *erytheme indure des scrofuleux*) is an interesting affection, not so uncommon as the others. I cannot do better than describe one of my own cases, which is typical of the milder form. The patient was a girl, aged twelve years, of unhealthy appearance and with enlarged cervical glands. The father had a history of tuberculosis. The skin of the lower extremities was of a livid color, cold and œdematous. Here and there, chiefly on the anterior aspects, were numerous nodules, some projecting above the surface, others only to be felt beneath it. Over the larger the skin showed a reddish tint and one had spread externally by a small punched-out mouth through which a thin pus was discharged. The disease was painless and indolent, with little tendency to healing. In graver cases multiple ulcers are formed, the openings of which may be connected by subcutaneous fistulæ. In my case the disease began at puberty. She recovered under tonic treatment and local antisepsis.

In conclusion, let me urge members of the profession to report the cases of cutaneous tuberculosis which they see in practice, of whatever kind. They are very few in dermatological clinics, and much light is needed upon the subject still, particularly in the case of the paratuberculoses.

HEREDITY IN PHTHISIS.

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On December 11, 1894, I read a paper before the Royal Medical and Chirurgical Society of London on "The Influence of Heredity in Phthisis," in which I compared the incidence of this disease on the children of phthisical parents with that on the children of non-phthisical parents living under similar conditions. In that paper I showed that in 275 families in which there was no phthisis in the parents, comprising a total of 1745 children, 24.87 per cent. of these became phthisical; while in 199 families of phthisical parents, with a total of 1182 children, 33.71 per cent. of the children became phthisical. This gives a difference of only about 9 per cent.